Diagnosis of Deafness in Preschool Children

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• **Hearing and speech disorders in children are often not due to actual defects in the ear or speech organs. Supposed loss of hearing and speech can occur in children who hear well but who cannot identify the words, understand their meaning or express them because of damage to certain brain centers and nerve pathways in conditions called aphasia, psychic disorders and mental deficiency.**

The child uses his special senses (hearing, sight, touch, feel and smell) to establish contact with his environment and of these senses, hearing is most important because it helps to build the best avenue of communication with his immediate world.

As the deafened child can usually be helped by treatment, it is necessary to make a correct diagnosis early in order that the proper measures may be instituted in the first five years of life.

THERE ARE SEVERAL clinical entities that cause hearing and speech disorders in children, and the treatment differs for each. As the proper remedial treatment in the formative years can mean much for a child's social and economic future, it is a physician's duty to make a correct diagnosis early.

The terms *hearing* and *deafness* are used in their functional sense in this communication. This means that normal hearing consists not only of an auditory stimulus passing from the peripheral ear to the area 41 in the brain, but also of its transmission to higher perceptive and psychologic stations where the sound is put to its destined purpose. In these interpretive centers the sound is memorized and stored for future symbolic use and is then passed on in its proper context for expression by the motor center for speech. The term *deafness* refers to a decrease in hearing function which may vary from slight to a total loss.

Diagnosing deafness in a preschool child is fraught with difficulties, not the least of which is determining whether the deafness is due to an auditory cause, to brain damage or to psychologic aberration. Often the child cannot be examined satisfactorily by pure tone or speech audiometry, and the other formal diagnostic procedures such as peep show tests, electroencephalography and psychogalvanic skin reaction studies require equipment and personnel not usually available.[1,2,5,6] Most of these procedures have their greatest value in estimating quantitative loss of hearing rather than in helping to make a differential diagnosis. Without formal hearing tests, the examiner must rely on the history and a careful observation of the child's behavior in relation to his environment.

It is necessary to keep in mind the normal growth of a child with regard to his mental, physical, social and emotional development so that this standard may be compared with the behavioral characteristics of children with hearing disorders.[7] The normal-hearing baby can be informally tested by simple procedures and serial observations as to how he reacts to sounds, words and gestures.[9] Ten to 30 days after birth, a sharp tone near the baby will usually evoke an auropalpebral reflex. After approximately six weeks of life the baby will stop voluntary movements of head, arms and legs when he hears a fairly loud noise. At from two to four months of age, infants begin turning the head toward the source of sound. Somewhat later there will be cooing, laughing and smiling, and the cry and jargon will have definite tonal inflections. Still later there will be gradual assimilation of ability to integrate sounds and gestures—enough so that adults can interpret their meaning—and at the same time the infant becomes increasingly aware of his environment. Simple single words normally are uttered at from 12 to 14 months of age, and by the 18th to 22nd month the child should be using sentences. The chronological succession of events just noted in the development of the normal baby depends upon unimpeded passage of the auditory stimuli through the multiple brain centers dealing with memory, symbolization and expression.

In this development process, the auditory messages are also integrated with those from other special senses to form a scaffold for the construction of language. The building blocks for language are (1) perception, (2) assimilation and interpretation, and (3) expression in correct context of sounds, words and gestures.[8] When language function is delayed or hampered for any reason, the child will have difficulty in communicating with his environment; and as a consequence his mental, social and behavioral development will be retarded and changed.

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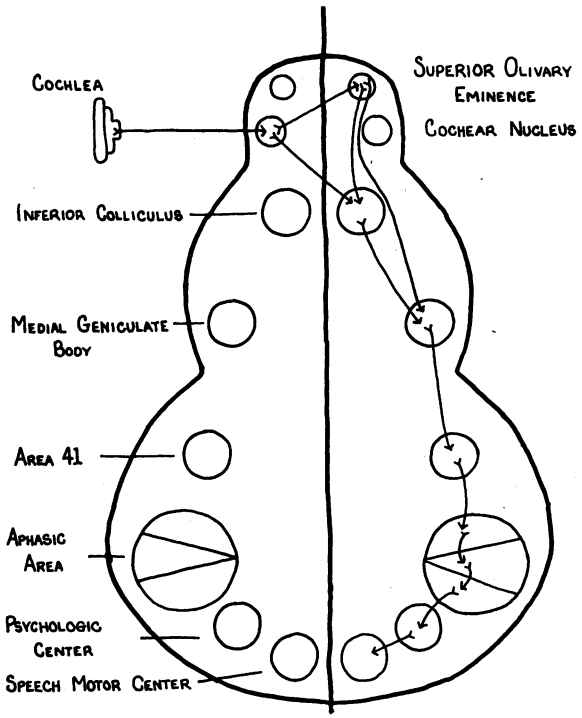

Figure 1

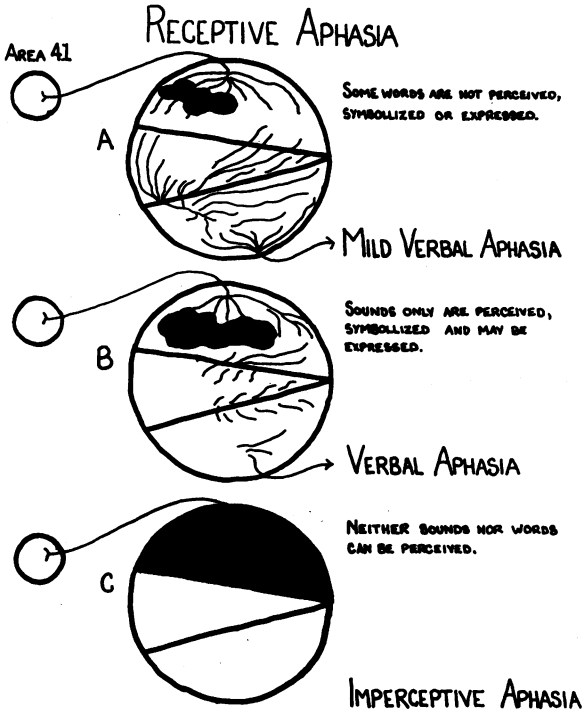

Figure 2

It is known that lack of language may be caused by many clinical entities. The following conditions have been found to comprise the majority of the hearing and speech disorders in children:

Peripheral Deafness. The causes are well known and discussion here is unnecessary.

Intermediate Deafness. Deafness due to lesions in the auditory pathway from the cochlear nucleus to the contralateral area 41 in the temporal lobe has been termed *intermediate deafness* (see Figure 1). This pathway is known to be vulnerable to trauma, as in birth injuries, and to lesions produced by intravascular hemolysis in Rh factor incompatibilities.[3,4] It seems that a more specific localization of the site of disease can be expressed by the term *intermediate deafness* rather than *central deafness*, a prevalent term. Use of the term *central deafness* should be limited for those cases with bilateral lesions in the areas 41.

Aphasia is a language disorder resulting from brain damage and it is characterized by poor symbolic behavior and psychologic aberrations. It may be mild or severe and there are different patterns of behavior dependent on how localized or extensive be the brain deterioration. When the auditory stimulus reaches area 41 in the temporal lobe, it is then transmitted to higher centers in the brain for more definitive interpretation. Normally the stimulus is processed in a receiving station (perception), then sent on to an integration center for memory and thinking to himself (symbolization) and finally reaches the expressive center where words or symbols are gathered together and transmitted in the correct context to the motor center for speech. In discussing the several types of aphasia it will be of help to consider several schematic drawings (Figures 2, 3 and 4).

Receptive auditory aphasia occurs when brain damage involves the perceiving center for sounds and words. If the damaged area be small and localized (Figure 2A) there will be minor impairment. Should the deterioration be greater, no words would be perceived (Figure 2B). When the damage is still more extensive, neither words nor sounds could be understood (Figure 2C). This is classed as *imperceptive auditory aphasia.* Children with receptive aphasia are unable to form language because they are confused as to the meaning of the sounds and words that they hear. They cannot segregate and focus attention on the words and sounds which should be of immediate importance to them. This confusion results in the child's speaking seldom, not using his voice purposefully, having a meaningless jargon and responding sluggishly to other sensory stimuli. Their social development is characterized by their inadequacy in playing with others and a lack of enthusiasm for the world in general, being neither shy nor aggressive. These children are easily distracted, are uninhibited and often persist in doing meaningless acts over and

over. Motor development is usually retarded and their coordination is poor.

Central auditory aphasia is produced when brain damage involves the central areas as shown in Figure 3A. Clinically a pure central aphasia is seldom found because the brain damage usually extends to the areas A and B. If brain deterioration is extensive in the central area, the characteristics of both receptive and expressive aphasia are found: The child fails to symbolize the words and sounds he has perceived, and as a result he is unable to think to himself. This acts as a severe retardant to development of language and as a group the children with this condition are the most retarded in social, behavioral and mental growth.

Expressive auditory aphasia occurs when a lesion damages the area shown in Figure 3B. The child is able to perceive and to symbolize internally but is unable to express the words in their correct context. As a group, children with such damage have milder emotional and social deviations from normal than the other aphasias because they are able to understand speech and to think to themselves, and are only handicapped by the inability to communicate by voice.

Psychogenic deafness is thought to be due to a defect in the brain area dealing with communications between the child and his environment. This region has not been localized anatomically but it is thought to be near the other cerebral centers (Figure 4). Because of emotional disturbances between the child's inner needs and the demands of his world, an impasse may develop in which he is unable to satisfy either. The children in this group have decided to give up their environment. The sense of hearing seems to be the more common link in communication with his environment to be abandoned, although visual stimuli may also be ignored. With the relinquishment of the sense of hearing the child becomes severely handicapped because of poor language foundation. Psychic deafness is thought to be generally defensive in nature and it is now recognized that a feeling of insecurity may be engendered by a lack of love and too frequent admonitions and punishments—the latter being significantly associated with the spoken voice. The auditory stimulus moves normally through the peripheral tracts to the higher cerebral centers where perception, internalization and expression are integrated. There is normal perception, internalization and the knowledge of how to express words. However, when the relay passes into the psychologic center the message is halted and does not go through to the motor center for the speech organs. This results in the suppression of speech which in turn causes deterioration in the child's general behavior with relation to his environment.

Infantile autism is known as a form of schizophrenia which develops in early infancy. Because of the psychic block, functional hearing and speech are not acquired and the baby's maturational growth is retarded. These children respond poorly to formal hearing tests and are often classed as being peripherally deaf. It has been noted that a large number of these babies are emotionally starved from lack of love and understanding. Unless the psychic barrier be removed, the child will never talk.

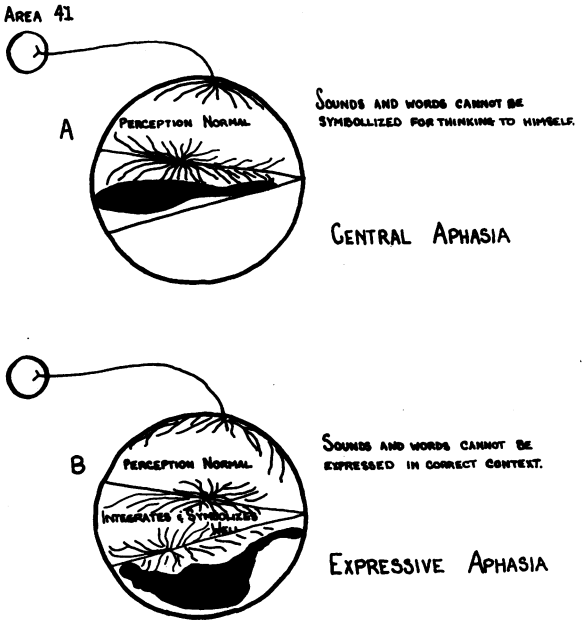

Figure 3

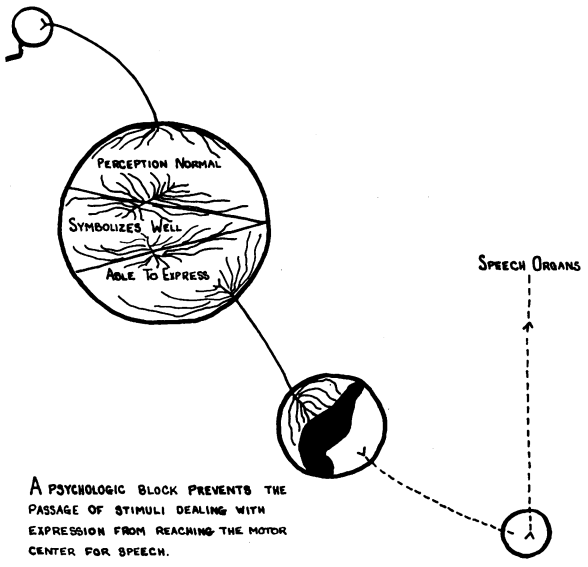

Figure 4

CHART 1.—*Points to Cover in History-Taking for Diagnosis of Childhood Deafness*

PRENATAL:
- German measles?
- Miscarriages, abortions or stillbirths?
- Unusual drugs during pregnancy?
- Accidents during pregnancy?
- Sickness during pregnancy?
- Length of pregnancy?

NATAL:
- Did labor begin normally or were drugs given?
- Was it difficult or long labor?
- Were forceps used?
- Was anesthesia given?

POSTNATAL:
- Was baby's skin yellow?
- Was baby's skin blue?
- Did baby have trouble nursing?
- What sicknesses has baby had?
- Has baby had high fevers? How high?
- Has baby had convulsions?
- Has baby had skull injuries? Been "knocked out"?
- What medicines has baby had?

FAMILY HISTORY:
- Any deafness in either mother's or father's family?
- Any deafness or no talking in baby's brothers or sisters?
- Any convulsions in family?
- Any mental disease in family?

BABY'S GROWTH:
1. When did baby first "hear" anything?
2. When did baby sit up alone?
3. When did baby creep?
4. When did baby stand alone?
5. When did baby walk?
6. When did child first talk?
7. Does child walk and run well?
8. Is child more awkward than other children?
9. When did child use sentences?
10. When was child toilet trained?
11. Does child play well with other children?

Schizophrenia usually is manifested after a year and a half to two years of age. Up to this point the child develops fairly normally in most respects and speech has been acquired before the psychic block occurred. In most cases one can elicit a history of pronounced fear of sounds from infancy. The child seems withdrawn from reality and exhibits bizarre relations with his environment. Even though he rejects sounds objectively, he does use them subjectively for his own amusement in phantasy form. It is characteristic that these children are the most mute of all, yet their motor development is nearly normal.

Obsession and anxiety neuroses are conditions in which hearing is relinquished only for certain situations. Generally speaking, children with psychic disorders of hearing will talk less, use and respond to gestures less and react to other stimuli less than will those with the other hearing disorders.

Mental deficiency may be confused with peripheral deafness, especially in children having an intelligence quotient from 45 to 65. In the case of lower intelligence quotient the motor, physical and behavioral characteristics are such that mental retardation is suspected and the apparent hearing loss can be accounted for. Many of these children in the 45 to 65 intelligence quotient bracket are being treated for auditory deafness. If their general inadequacy in most situations is recognized, then their slow speech development can be predicted. If the intelligence quotient is 50, then the child will have a mental age of one when he is two years old and his speech formation and other phases of development will take twice as long as the normal child.

CHART 2.—*Occurrence of Various Phenomena in Several Categories of Childhood Deafness*

Phenomenon	Peripheral Deafness	Aphasic Deafness	Psychic Deafness	Mental Deficiency
Hears loud sounds usually	X	....	....	X
Hears loud sounds now and then	....	X	X	....
Hears sounds but not speech	....	....	X	....
Doesn't want to hear	....	....	X	....
Afraid of loud sounds	....	....	X	....
Confused by words and sounds	....	X	....	X
Uses what hearing he has to his advantage	X	....	....	X
Doesn't use hearing to his advantage	....	X	X	....
Listens well	X	....	....	....
Listens poorly	....	X	X	....
Does he talk some?	X	X	....	X
No speech	....	....	X	....
Talked and then stopped talking?	X	....	X	....
Talking delayed after 2 years	....	....	....	X
Uses voice to call people or make them notice him	X	....	....	X
Voice tones are normal tones	....	X	X	X
Voice tones are not normal tones	X	....	....	....
Voice used in playing	....	X	....	X
Voice not used in playing	X	....	X	....
Uses and understands gestures and facial expressions	X	....	....	X
Doesn't use or understand gestures	....	X	X	....
Does he use eyes well and sense of touch?	X	....	....	....
Poor use of eyes and sense of touch	....	X	X	X
Sit, creep, walk, run, etc., normal	X	....	X	....
Awkward or poor movements	....	X	....	X
Is active and accomplishes things	X	....	....	....
Is active and doesn't accomplish much	....	X	....	X
Shuffles and balance is unsteady	X	....	....	....
Coordination is good	X	....	X	....
Coordination poor	....	X	....	X
Laugh and cry have "flat" tones	X	....	....	....
Laughs and cries without reason	....	....	X	....
Doesn't laugh or cry very loud or easily	X	X	....	X
Laugh and smile to themselves only	....	....	X	....
Does things over and over again	....	X	....	....
Doesn't do anything for very long	....	X	....	....
Easily disturbed	....	X	....	....
Likes people around him	X	....	....	X
Doesn't care if people are around	....	X	X	....
Temper tantrums	....	X	X	....
Dislikes loving and cuddling	....	....	X	....
Doesn't play well with other children	....	X	....	....
Gets along well with people	X	....	....	X
Doesn't care for other people	....	X	X	....

(X) Indicates Conditions in Which Abnormality Occurs

Other conditions resulting in impaired speech are *dysarthria* and *anarthria*. The site of pathologic change is the motor speech center and therefore the speech impairment is from motor deprivation rather than sensory loss as in aphasia. The speech center lesion is caused by brain deterioration which often is associated with aphasia, mental deficiency and birth injuries. In addition to the speech impairment there is a dysfunction of chewing and swallowing, these being allied functions of the speech muscles. This deglutitory deficiency serves to differentiate dysarthria from expressive auditory aphasia, which has no motor disturbance.

It is of considerable help in the taking of a history and in the observation of the child's behavior to have a planned approach. Use of the forms shown in Charts 1 and 2 has proven of value in avoiding the possibility of failing to ask necessary leading questions. When the results are tabulated a clearer insight often can be gained and further definitive questions can be asked to aid in making a correct differential diagnosis.

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REFERENCES

1. Bordley, J. E., and Hardy, W. J.: A study in objective audiometry with use of psychogalvanometric response, Ann. Otol., Rhin. & Laryng., 58:751-760, Sept. 1949.

2. Dix, M. R., and Hallpike, C. S.: The peep show: A new technique for pure tone audiometry in young children, Brit. J. F., 2:719, Nov. 8, 1947.

3. Dublin, W. B.: The neurologic lesion of erythroblastosis fetalis in relation to nuclear deafness, Am. J. Clin. Path., 21:935-939, Oct. 1951.

4. Goodhill, V.: The nerve deaf child: Significance of Rh, maternal rubella and other etiologic factors, Ann. Otol., Rhin. & Laryng., 59:1123-1147, Dec. 1950.

5. Guilford, F. R., and Haug, C. O.: Diagnosis of deafness in the very young child, Arch. Otol., 55:101-106, Feb. 1952.

6. Marcus, R. E., Gibbs, E. L., and Gibbs, F. A.: Electroencephalography in the diagnosis of hearing loss in the very young child, Dis. Nerv. System, 10:170, June 1949.

7. Myklebust, H. R.: Auditory Disorders in Children, Grune & Stratton, Inc., New York City, 1954.

8. Myklebust, H. R.: Changing concepts in audiology, Laryng., 66:437-444, April 1956.

9. Wishart, D. E. S.: The hard of hearing infant, Ann. Otol., Rhin. & Laryng., 63:378-393, June 1954.